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CASE REPORT

Adult idiopathic hypertrophic pyloric stenosis

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Idiopathic hypertrophic pyloric stenosis (IHPS) is a predominantly infantile disease. The adult type of IHPS is extremely rare but it has been well recognized since the 19th century. We report a case of a 47-year-old male patient who presented with postprandial nausea and vomiting. He underwent upper gastrointestinal endoscopy, and gastric outlet obstruction was discovered. The upper gastrointestinal barium study showed a distended stomach with delayed gastric emptying due to pyloric stenosis. The abdominal computed tomography revealed thickening of the distal stomach. Since gastric malignancy could not be excluded, he underwent antrectomy with Billroth I anastomosis. The pathology revealed no malignancy but showed hypertrophy and hyperplasia of the inner circular muscle of the pylorus, which was compatible with IHPS. We reported the case to remind young physicians of this rare disease.

Introduction

Idiopathic hypertrophic pyloric stenosis (IHPS) is a predominantly infantile disease, whose incidence is between 0.1% and 0.8%.[1] Infantile IHPS is always diagnosed and treated during the first 2 months of life. The adult type of IHPS is so rare that most physicians nowadays are not aware of it. In the French atlas of pathologic anatomy *Anatomie pathologique du corps humain*, published in 1842, Cruveilhier first described the pathologic anatomy of adult IHPS.[2] Maier confirmed it as a clinical disease entity based on a series of pathologic findings at necropathy in 1885.[3] However, there are less than 300 cases reported in the literature.[4–6] We report a case of adult IHPS to remind young physicians of this rare disease.

Case report

The patient is a 47-year-old male Taiwanese. He complained of epigastric fullness and easy satiety for 1 month. The symptoms persisted and worsened, leading to

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postprandial nausea and vomiting lately. He had no specific medical or surgical history. He had no history of peptic ulcer disease or diabetes mellitus. He visited our outpatient department and underwent upper gastrointestinal (UGI) endoscopy. The endoscopy showed a distended stomach with a constricted pylorus (Fig. 1). The scope passed the pylorus with slight resistance, and mild superficial erosions at the area of narrowing were noted. He was admitted under the diagnosis of gastric outlet obstruction. Physical examination revealed a soft and flat abdomen. No tenderness or palpable mass was detected. He had no leukocytosis or anemia. He underwent UGI barium study, which showed delayed emptying of the stomach and an elongated narrowing pyloric channel (Fig. 2). Abdominal computed tomography (CT) was performed. Thickening of the wall in distal stomach was noted (Fig. 3). Because gastric malignancy could not be excluded and the symptoms persisted, he underwent surgery. The pylorus was thickened and no adhesion was noted in laparotomy. He underwent antrectomy with Billroth I reconstruction. The surgery was uneventful. The pathology revealed no malignancy but showed thickening of the pyloric muscle (Fig. 4). Prominent hypertrophy and hyperplasia of the inner circular muscle layer of pylorus were noted. Hypertrophy of muscularis mucosae with muscle strands within the lamina propria was also noted. The pathologic picture was consistent with IHPS in adults. His symptoms were much improved after surgery.

Discussion

Hypertrophic pyloric stenosis (HPS) could be classified as primary and secondary. The secondary type are more common and may be induced by many causes, including healing of previous gastric or duodenal ulcers, hypertrophic gastritis, carcinomas, gastrointestinal stromal tumors, bezoars, vagal hyperactivity, and postoperative extrinsic adhesions.[6–8] This type of HPS has no or only mild hypertrophy of the pyloric muscle. The muscle fibers are usually replaced by fibrous tissues. Development of HPS without predisposing factor is defined as primary or idiopathic type, and is characterized by hypertrophy and hyperplasia of pyloric muscle. IHPS is always diagnosed in infants, and the adult variant of IHPS is extremely rare. The exact occurrence of adult IHPS cannot be estimated accurately, since some of the cases may remain asymptomatic. With no known reason, adult IHPS occurs more commonly among middle-age males,[9,10] as is the case with the patient in this report.

Figure 1 The stomach is filled with much undigested food residues. The pylorus is constricted and had no relaxation during examination.

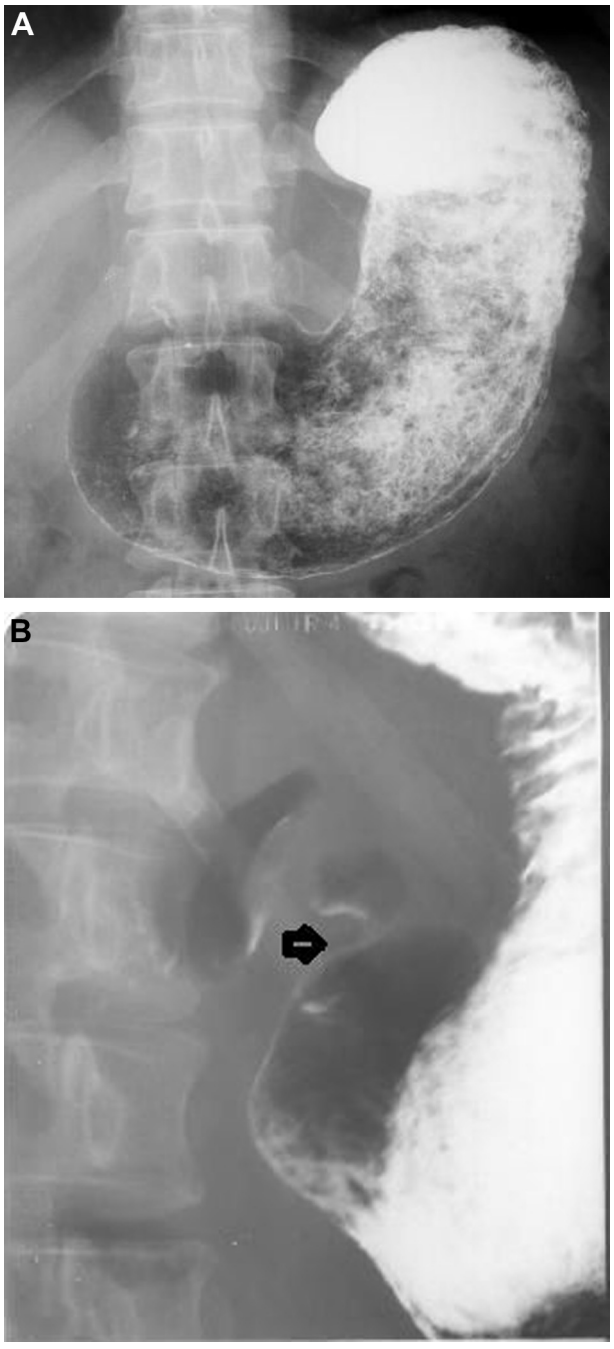

Figure 2 (A) The UGI series shows a marked distended stomach filled with much food debris. (B) The pylorus is elongated and narrowed (arrow).

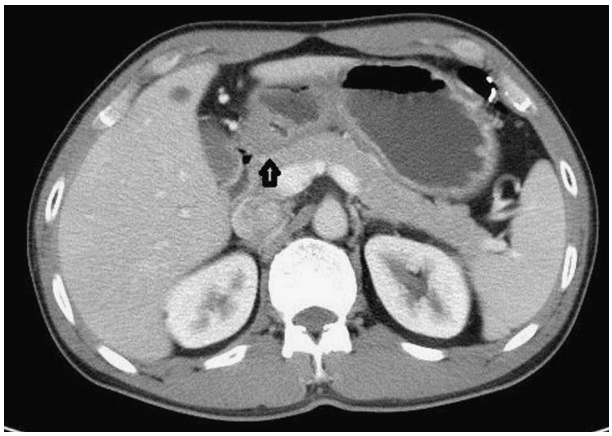

Figure 3 CT scan revealed thickening of the distal stomach (arrow).

The etiology of IHPS is still uncertain. It seems that both genetic and environmental factors are involved. Most physicians believe that adult IHPS is due to the persistence of the mild infantile form that persists into adult life.[8,11–13] In fact, infantile and adult IHPS have similar anatomical and histological changes.[14] Hypertrophy of the pylorus may not become symptomatic until some predisposing factors such as edema, spasm, or inflammation precipitate pyloric occlusion have occurred.[15,16] Other possible etiologies that have been proposed are protracted pylorospasm, vagal hyperactivity, and neuromuscular incoordination due to changes in Auerbach's plexus.[4–16] Several authors have reported a familial tendency of occurrence of infantile and adult IHPS, and both forms can occur in the same family.[17,18] However, most cases of adult IHPS appeared to arise *de novo*. Cockayne and Penrose[19] concluded that there is a definite although small familial tendency.

The clinical symptoms of adult IHPS are similar to those of gastric outlet obstruction induced by other causes, including epigastralgia, easy satiety, and postprandial nausea or vomiting. Unlike infantile IHPS, an abdominal mass is seldom felt in patients afflicted with adult IHPS.

Diagnosis of adult IHPS before surgery is not easy. Radiology examination usually shows no pathognomic findings and

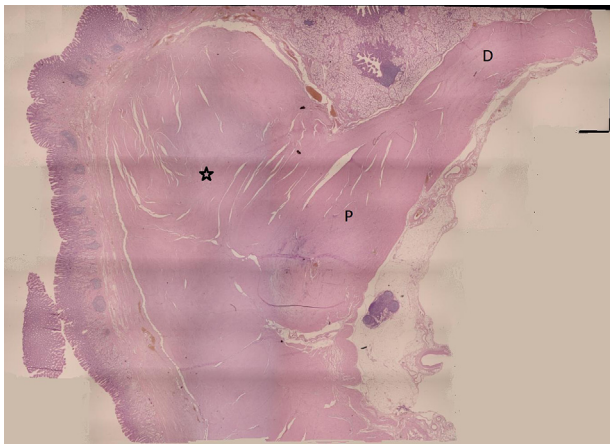

Figure 4 Cross section of the pylorus shows thickening of the inner circular muscle layer of the pylorus (star). D = duodenum; P = pylorus (10× H&E).

can be normal in some cases.[13] Usually, the UGI series will show delayed gastric emptying due to pyloric stenosis. The pyloric canal is usually elongated and narrowed. A mushroom-like deformity is used to describe the bulging of the pyloric muscle into the duodenal cap. Kirklin and Harris[11] were the first to define the classic radiological findings in 1933. This feature, however, may be produced in normal patients by manual pressure on the stomach.[16] UGI endoscopy is needed to rule out other causes of gastric outlet obstruction. The typical endoscopic picture of adult IHPS is a fixed, markedly narrowed pylorus with a smooth border. Schuster and Smith[20] had defined this unique endoscopic sign as "cervix sign." Transabdominal sonography is not so useful in the diagnosis of adult IHPS compared to infantile IHPS, although some had advocated its usefulness.[20] Abdominal CT scan is helpful to exclude the secondary type of HPS. Some cases of adult IHPS showed thickening of the distal gastric wall in CT scan, like our case did, but it was not specific. Retrospectively, we may find that the thickening of the distal stomach in our case is actually attributable to hypertrophic pylorus. Maybe this could be a useful diagnostic criterion in the future since the resolution of modern-day CT is much better than before. Theoretically, endoscopic ultrasonography will have its advantage in diagnosing the rare disease. Nonetheless, a definite diagnosis is usually made by a pathologist. Grossly, the most significant finding is the elongated and thickened pylorus, which can be the first impression of the surgeon during laparotomy. The normal thickness of the muscle of the adult pyloric canal ranges from 3 to 8 mm with an average of 4 mm.[8,12,21] In adult IHPS, it is increased to 1–1.5 cm on average, and measurements of up to 3 cm have been recorded.[9] It is 1.2 cm in our case. Microscopic examination will demonstrate marked hypertrophy and hyperplasia of inner circular muscle layer of pylorus, which may be associated with reactive mucosal gastropathy.[8,13] The presence of fibrosis in addition to smooth muscle hypertrophy can often be present. However, there should be no process of inflammation or neoplasm seen in the muscularis propria.

Surgery is indicated in the treatment of adult IHPS. Partial gastrectomy, gastroenterostomy, pyloromyotomy, and pyloroplasty have been proposed as treatments for adult IHPS by various physicians.[6,8,15,16,22,23] Endoscopic dilatation had also been advocated but it has a high recurrence rate.[8,24] Some authors prefer partial gastrectomy because carcinoma may be a complication of long-standing pyloric hypertrophy.[6]

Conclusion

Adult IHPS is an ancient disease although its etiology is not clear. We should be aware of this rare disease. Treatment with partial gastrectomy with Billroth I reconstruction is preferred by most physicians.

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